Incidence of Odontogenic Keratocysts in Patients with Gorlin-Goltz Syndrome according to Age, Gender and Location

Božidar Pavelić[1]
Kruna Valter[2]
Vanja Vučićević-Boras[3]
Davor Katanec[4]
Sonja Levanat[5]
Karl Donath[6]

[1]Department of Dental Pathology
School of Dental Medicine
University of Zagreb
[2]Private Dental Practice, Zagreb
[3]Department of Oral Medicine
School of Dental Medicine
University of Zagreb
[4]Department of Oral Surgery
School of Dental Medicine
University of Zagreb
[5]Department of Molecular Medicine
Institute "Ruđer Bošković", Zagreb, Croatia.
[6]Institute of Oral Pathology
University of Hamburg, Germany

Summary

The aim of this study was to determine the appearance of odontogenic keratocysts (OKC) and keratocyst-like lesions (KLL) in patients with Gorlin-Goltz syndrome, according to age and gender of participants, and in relation to the place of origin (mandible, maxilla, soft tissues, which include the mucosa of the alveolar ridge, skin of the nose and face, parotid gland together with maxillary sinuses). From 1965 until 1998, 58 OKC were found in 30 patients with Gorlin-Goltz syndrome and confirmed by histopathologic analysis. Patients were aged from 10 to 90 years. Average age of the patients with Gorlin-Goltz syndrome was 33.71 years. Average age in males was 35.53 years and in females 31.12 years. OKC connected with Gorlin-Goltz syndrome were more frequently found in males (58.62%) compared to the females (41.38%). The peak incidence of OKC in patients with Gorlin-Goltz syndrome was between ages 21-30 years. According to the location, OKC were found predominantly in the mandible (60.34%), the maxilla (15.52%), soft tissues (13.79%), and in maxillary sinuses (10.34%).

Key words: *odontogenic keratocysts, Gorlin-Goltz syndrome.*

Acta Stomat Croat
2004; 23-25

ORIGINAL SCIENTIFIC PAPER
Received: January 3, 2004

Address for correspondence:

Božidar Pavelić
Department of Dental Pathology
School of Dental Medicine
Gundulićeva 5, 10000 Zagreb
Croatia
tel: 4802 126/136
fax: 4802 159
e-mail:
bozidar.pavelic@zg.hinet.hr

Introduction

Odontogenic keratocysts (OKC) are classified as developmental epithelial cysts which comprise approximately 11% of all cysts of the jaws (1, 2). One of the most characteristic features of OKC is the high recurrence rate(3). OKC may occur as a sporadic OKC or together with Gorlin-Goltz syndrome (nevoid basal cell carcinoma syndrome). Gorlin-Goltz syndrome is characterized by multiple basal cell carcinomas, odontogenic keratocysts of the jaws, palmar and/or plantar pits, skeletal anomalies (most frequently seen on vertebral bones and ribs), and intracranial calcifications (4-6). Gorlin-Goltz syndrome is an autosomal dominant inheritance disorder, and probably a result of abnormalities located on the chromosomes 9 (7). Recently it was reported that the loss of human patched (ptch), a tumor suppressor gene, could be the possible molecular origin of OKC (8). Most frequently it becomes apparent during the second or third decade of life (9). OKC are usually the first sign of Gorlin-Goltz syndrome, and can occur also in patients under the age of ten. It has been hypothesized that OKC are frequently found earlier in patients with Gorlin-Goltz syndrome compared to sporadic OKC (10). In one patient, 1-30 cysts may develop (11). Therapy may include various types of surgical procedures, often with high recurrence rate or some other early postoperative complication (12, 13). The most effective way to follow-up the bone healing and tissue density after surgical procedure is by radiovisographical (RVG) densitometry (14, 15).

Materials and methods

In this study, 30 patients with 58 OKC and Gorlin-Goltz syndrome were included. The patients were recruited from the Institute of Oral Pathology in Hamburg, Germany and from the School of Dental Medicine (Department of Dental Pathology, Oral Medicine, Pediatric Dentistry and Oral Surgery) in Zagreb, Croatia, between the years 1965-1998. All the OKC were confirmed histopathologically. They were subdivided according to the age and gender of the participants and location in the orofacial system. According to the location OKC were divided into four groups: in the mandible (anteriorly and posteriorly), in the maxilla (anteriorly and posteriorly), in soft tissues which comprises the mucosa of the alveolar ridge, parotid gland, skin of the nose and face, and in the maxillary sinus (either left or right). OKC located anteriorly were those situated between canines, and those located posteriorly were situated distal to the first premolars.

Results

Age and gender

The patients included in this study were aged from 10 to 90 years. The average age of patients with Gorlin-Goltz syndrome at the time when OKC were diagnosed was 33.71 years. In males the average age was 35.53 years and in females 31.12 years. Out of 58 diagnosed OKC, 34 (58.62%) were found in males and 24 (41.38%) in females. Male to female ratio was 1.4:1.

The highest incidence of OKC in all patients with Gorlin-Goltz syndrome was determined between ages 21-30 years (Table 1).

Location

Out of 58 diagnosed OKC in patients with Gorlin-Goltz syndrome, 35 (60.34%) were located in the mandible, 9 (15.52%) in the maxilla. In the mandible, two OKC were situated anteriorly, and 33 (94.29%) posteriorly. In the maxilla, one OKC was found anteriorly, and 8 (88.89%) were found posteriorly. 8 (13.79%) KLL were found in the soft tissues which include mucosa of the alveolar ridge, parotid gland, skin of the nose and face. Six (10.34%) OKC were detected in the maxillary sinus, with unequal distribution of one in the left sinus and 5 (83.83%) in the right sinus (Table 2).

Discussion

Odontogenic keratocysts probably arise from any of the primordial epithelia (the dental lamina or its remains) or, as originally believed, from the enamel organ before tooth formation. The histological appearances of the lining of keratocysts are characteristic: a flat basement membrane, elongated palisated basal cells, corrugated parakeratotic surface,

with or without keratin layer in the lumen. Most cysts in the orofacial region, with keratin in the lumen, were often diagnosed as keratocysts, which lead to misunderstanding and consequently wrong diagnosis. Thus these lesions are similar to OKC, but they are not the same. Therefore, in this study all the cysts found in soft tissues were called keratocyst like lesions.

Sabbia et al. (13) and Addessi et al. (14) reported that in 90% of patients, nevoid basal cell carcinoma syndrome is associated with recurring OKC. Lo Muzio et al. (5) reported that OKC were the first symptom of Gorlin-Goltz syndrome in 78% of their cases. The same authors also underlined that OKC occur earlier in patients who have nevoid basal cell carcinoma syndrome when compared to the patients who do not have the above mentioned syndrome.

Previous investigations showed that connection between OKC and Gorlin-Goltz syndrome is higher than 5%. Payne (15) reported that the incidence of OKC together with Gorlin-Goltz syndrome is 7.4%, Radden and Reade (16) found connection between OKC and Gorlin-Goltz syndrome in 6% of patients and Hodgkinson (17) in 8.8% of studied cases. Lam and Chan (18) reported that multiple cysts occurred in 9% out of 69 ethnic Hong Kong Chinese patients, and only three cases were diagnosed as Gorlin-Goltz syndrome. Oda et al. (19) found Gorlin-Goltz syndrome in 5% of their patients with OKC, with a higher distribution in the first and second decades. Results of our study show that the higher distribution of OKC together with Gorlin-Goltz syndrome occured during the second and third decade, but also with a second peak during the sixth decade of life in the studied patients. Dowling et al. (20) stated that although the occurrence of OKC in Gorlin-Goltz syndrome is usually during the second or third decades of life, it should be noted that it can occur earlier as seen in their patient, who was 5 years old, which was the first manifestation of nevoid basal cell carcinoma. Lo Muzio et al.(10) found that the youngest patient with OKC and Gorlin-Goltz syndrome was eight years old. Results of our study show that the youngest patient with OKC together with Gorlin-Goltz syndrome was ten years old.

According to gender, male preponderance was found in this study and male to female ratio was 1.4 : 1.

We also found that number of OKC in every patient with Gorlin-Goltz syndrome varied a lot. Seventeen patients had one OKC together with Gorlin-Goltz syndrome, and nine patients had two OKC at the same time. In one patient, 12 OKC were diagnosed during the period of 12 years, and two OKC occurred simultaneously four times. In two patients, three OKC were diagnosed at the same time. In another patient in whom 5 OKC were found, two of them were diagnosed at the same time on the left side of the mandible and KLL on the skin of the nose. On the basis of these data we can conclude that in patients with Gorlin-Goltz syndrome, multiple OKC occur frequently at the same time, or that they can be diagnosed simultaneously. Therefore, in every patient in whom one OKC is diagnosed, it is reasonable to perform additional investigations in order to exclude coexistence of another OKC.

This study also highlights the need to consider Gorlin-Goltz syndrome as a possible diagnosis in all patients with odontogenic keratocyst. Last, but not least, in every patient with Gorlin-Goltz syndrome it is advisable to make further familial investigations.